Atypical unfamiliar face processing in Williams syndrome: What can it tell us about typical familiarity effects?

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Abstract

Introduction: Familiar and unfamiliar face perception is typically dissociated by the relative use of internal and external face features. The Williams syndrome (WS) social phenotype emphasises hyper-sociability, with an interest in interacting with people irrespective of familiarity. The aim is to explore whether unfamiliar face processing is characterised by the typical dissociation between internal and external features in WS, or whether the social stimulus drive towards strangers is linked to atypicalities of unfamiliar face processing. **Method:** The procedure replicates that previously used with typically developing children. Participants with WS (aged 10-18 years) and typically developing comparison participants determine whether two face parts are from the same person or different people, using the whole face, internal and external features. **Results:** Only participants with WS, and not typically developing participants, show greater accuracy matching unfamiliar faces from internal than external features. **Conclusions:** Evidence of atypical unfamiliar face processing in WS may inform models of typical face perception, revealing the origins of the relative advantage for internal features typically associated with familiar but not unfamiliar faces. The results also have implications for understanding more clearly the social phenotype associated with WS.

Abbreviations: BPVS: British Picture Vocabulary Scale; WS: Williams syndrome; RCPM: Ravens Coloured Progressive Matches

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Williams syndrome (WS) is a genetic disorder arising from a micro-deletion of approximately 25 genes at chromosome site 7q11.23 (see Donnai & Karmiloff-Smith, 2000). Previous research has mapped the distinct neuropsychological profile of WS showing strengths and weaknesses of cognitive ability (Bellugi, Lichtenberger, Jones, Lai & St George, 2000). As well as differences between domains (verbal-nonverbal), within-domain dissociations occur; spatial cognition deficits co-occur with more proficient face recognition (e.g. Bellugi et al., 2000). Related to the social importance of faces, a prime characteristic of WS is a high level of sociability.

Individuals with WS are characterised as overfriendly and unreserved (Gosch & Pankau, 1994) which has led to suggestions of a strong ‘compulsion’ towards social engagement (Frigerio et al., 2006). Similarly, subtle atypicalities of social behaviour, for example extended face gaze and a desire for social contact, have led to descriptions of ‘hyper-sociability’ (Jones et al., 2000). This is not confined to familiar people, as shown by abnormally high ratings of approachability for unfamiliar faces (Bellugi, Adolphs, Cassady & Chiles, 1999a; Frigerio et al., 2006) and parental reports of increased likelihood to approach strangers (Doyle, Bellugi, Korenburg & Graham, 2004). Empirical evidence suggests abnormally intense looking towards strangers by infants with WS (Mervis et al., 2003) and reduced face-disengagement in cognitively demanding situations (Doherty-Sneddon, Riby, Calderwood & Ainsworth, submitted). It is therefore argued that individuals with WS are highly, and often inappropriately, sociable with people irrespective of familiarity.

The human face captures attention (Vuilleumier, 2000) and must be processed successfully to allow smooth running interactions; not only must we identify friends but interpret a range of conversational signals. Individuals with WS are relatively good at interpreting facial cues; reading simple or complex emotions (Tager-Flusberg, Boshart & Baron-Cohen, 1998) and using eye gaze cues to infer intentions (Karmiloff-Smith, Klima, Bellugi & Grant, 1995). These two specific skills dissociate individuals with WS from those with autism (Riby, Doherty-Sneddon & Bruce, 2004) and it is clear that these two groups exhibit contrasting social phenotypes and face processing skills (cf. Brock, Einav, & Riby, in press). Due to a desire to engage with both familiar and unfamiliar people, the effect of familiarity on face perception may be particularly important when exploring social cognition in WS.

One paradigm employed to investigate the effect of familiarity on face perception contrasts the use of internal and external face features. Over the last two decades researchers have used methods of covering, cropping and blurring different face regions to identify the type of face features and information that are more or less useful for processing familiar and unfamiliar faces. Research with typical adults consistently shows that masking internal features (eyes, nose, mouth) of familiar faces is more detrimental for recognition and face matching than masking external features (hair, chin, ears; Ellis, Shepherd & Davies, 1979; Young, Hay, McWeeny, Flude & Ellis, 1985). This pattern is not evident for unfamiliar faces and there is no change in the way that internal and external features contribute to unfamiliar face processing across development (Campbell, Walker & Baron-Cohen, 1995; Campbell et al., 1999, Bonner & Burton, 2004).

The relative advantage for internal features of familiar but not unfamiliar faces has implications for face perception models. A popular theoretical model (Bruce & Young, 1986) suggests that familiar and unfamiliar faces are processed via independent routes, relying upon different functional codes. Familiar faces activate face recognition units (FRU) and person identity nodes (PIN) whereas unfamiliar faces proceeds through early structural representations of the face (via directed visual processing) where information is strategically encoded. The independence of strategies is supported both empirically and theoretically.

Given an interest in people irrespective of familiarity, the internal versus external paradigm may reveal facets of face perception in WS. An abundance of previous research has already shown that individuals with WS may not process facial identity using typical strategies (e.g.

Karmiloff-Smith et al., 2004; Deruelle, Mancini, Livet, Cassé-Perrot, & de Schonen, 1999; but see Tager-Flusberg, Plesa-Skwerer, Faja & Joseph, 2003) and show atypical brain activation (e.g. Grice et al., 2001; Mills, Alvarez, St. George, Appelbaum, Bellugi, & Neville, 2001). Therefore atypicalities of this nature may extend to explorations of both familiar and unfamiliar face perception. Specifically relating to the use of internal and external face features, previous research has shown that participants with WS (n=11, 5-17 years) are more accurate matching face parts (internal/external) to a whole face target using external than internal features (Deruelle, Rondon, Mancini & Livet, 2003). The same pattern was also found for typically developing participants on the same task (matched for chronological or mental age). However, high performance across conditions (due to matching to a whole face target) may have masked subtle group differences. Deruelle and colleagues concluded that individuals with WS use typical face processing strategies and perform at a level predicted by their chronological age. This contrasts with evidence from numerous investigations showing performance below chronological age level across face processing demands (e.g. Karmiloff-Smith et al., 2004; Riby, Doherty-Sneddon, & Bruce, 2004; in press).

The current task uses the internal versus external feature paradigm to explore face matching by individuals with WS replicating a procedure already used with typically developing individuals (Bonner & Burton, 2004). Whole face targets are eliminated and the stimuli incorporate different orientations to avoid pattern matching strategies. It is predicted that participants with WS will show atypical unfamiliar face matching linked to the social phenotype of the disorder. As individuals with WS exhibit a lack of distinction between familiar and unfamiliar people, evidenced by their abnormally high ratings of approachability for unfamiliar faces and extended face gaze towards strangers, it is hypothesised that this group will show a different pattern of internal and external feature use than typically developing individuals. The task specifically investigates unfamiliar face matching and predicts that participants with WS will show a pattern of performance more closely linked to familiar face processing than typically developing individuals on the same task. The relative use of internal features will be of particular interest and it is predicted that individuals with WS will show relatively more use of internal compared to external features than typically developing participants.

Method

Participants

Thirteen individuals with Williams syndrome[1] ranging from 10 years 2 months to 18 years 2 months participated (mean 13 years 6 months, SD 34 months; 8 male, 5 female). All participants were recruited through the Williams syndrome Foundation and consent was received prior to participation. Eleven participants were diagnosed with the fluorescent in situ hybridisation test, whilst two were diagnosed by clinicians based on the cognitive and behavioural characteristics associated with WS. Verbal ability was assessed using the British Picture Vocabulary Scale (2nd Edition, BPVS II; Dunn, Dunn, Whetton & Burley, 1997), providing a mean verbal mental age of 11 years 1 month (ranging 9 years 3 months to 13 years 8 months, SD 23 months). Nonverbal ability was assessed using the Raven's Coloured Progressive Matrices (RCPM; Raven, Court & Raven, 1990), providing a mean score of 15 (ranging 9 to 26, SD 6; max. 36).

Each participant with WS was individually matched to three typically developing participants: one of comparable verbal ability (VMA); one of comparable nonverbal ability, to account for the uneven profile of cognitive skills (NVMA); and one of comparable chronological age, to control for life-experience (CA). Using the BPVS II the VMA group had a mean verbal age of 11 years 2 months (ranging 9 years 1 month to 13 years 11 months, SD 24 months) with no difference between groups in this measure (p=.42). The VMA group had a mean

[1] This sample size is comparable to published research with this population, e.g. Deruelle et al. (2003), Karmiloff-Smith et al. (2004). Due to the rarity of WS and the size of the UK many published research studies use samples of this size.

chronological age of 10 years 8 months (ranging 9 years 0 month to 14 years 1 month, SD 25 months). The NVMA group had a mean RCPM score of 15 (range 9 to 26, SD 6) and a mean chronological age of 9 years 1 month (ranging 7 years 2 months to 11 years 4 months, SD 30 months). There was no difference in nonverbal ability across groups (p=.72). The CA group ranged from 10 years 0 months to 18 years 0 months (mean 13 years 5 months, SD 34 months), the groups did not differ in chronological age (p=.40).

Design and Procedure

All faces were unfamiliar to participants and were pictures of individuals who had previously given consent for the images to be used in research[2]. Eight faces were used and 'same' trials constituted a full-view and a ¾-view of the same person. 'Different' trials comprised a full- and ¾-view of different individuals of the same gender and ethnicity. The task comprised 24 trials, 8 in each condition (internal, external, whole) in randomized order and all participants completed all conditions. Removal of internal features from whole face stimuli was standardised using an oval shape encompassing eyes, nose and mouth. External features comprised the hair, ears, chin and face contour (Figure 1). All images were transformed to grey-scale and standardized to 200 x 300 pixels using image software. Each trial consisted of face pairs presented side by side (Figure 1). The stimuli replicated the exact parameters employed by Bonner and Burton (2004).

(Figure One)

[2] It would be interesting to incorporate familiar faces; however, it was not possible to obtain the same personally familiar faces for all participants. Previous research has used famous faces but it has been emphasized that these lack ecological validity due to an absence of interpersonal contact. Additionally, a number of parents noted that their child lacked interest in films or television and therefore even obtaining suitable famous faces would be problematic for individuals with WS.

Participants were tested individually at home or school. As a practice trial, participants were shown pictures of the experimenter taken from two different angles and told they were different views of the same person. For experimental trials, the participant decided if two images were of the same person or different people. Trials were presented on A4 size paper and placed in front of the participant until a verbal response was provided, thus being self-paced[3].

Results

A 3 x 4 analysis of variance (ANOVA) with factors Part (internal, external, whole) and Group (WS, VMA, NVMA, CA) showed a significant difference in accuracy between Groups $F(3,48)=19.87$, $MSE=211.18$, $p<.001$. There was no difference between the WS and VMA groups ($p=.93$; overall mean WS 81%, VMA 83%) however participants with WS performed more accurately than those matched for nonverbal ability (mean 69%; $t(12)=3.08$, $MSE=3.87$, $p<.01$) and less accurately than those of comparable chronological age (mean 94%; $t(12)=4.10$, $MSE=3.28$, $p<.01$), see Figure 2. The chronological age matched group performed significantly below ceiling level across conditions (compared to 100% $t(12)=6.16$, $MSE=2.29$, $p<.01$). However, taking the three trial conditions separately reveals that the CA group performed at ceiling for the whole face matching trials (compared to 100% $p=.17$), but importantly did not perform at ceiling for the internal trials ($t(12)=7.56$, $MSE=2.77$, $p<.001$) or the external feature trials ($t(12)=4.13$, $MSE=2.02$, $p<.05$).

(Figure Two)

[3] Reaction time analyses were not included here as they may have been particularly unreliable in the individualised testing sessions required for participants with WS. They would also have been particularly uninformative when accuracy was low. However, future research may explore this issue.

Overall, participants were significantly affected by the part of the face used for matching $F(2,96)=37.07$, $MSE=77.29$, $p<.001$. Whole face matching (mean 89%) was significantly more accurate than using internal (mean 75%; $t(51)=8.65$, $MSE=1.71$, $p<.001$) and external features (mean 82%; $t(51)=5.04$, $MSE=1.73$, $p<.001$). Additionally, external feature matching was more accurate than using internal features ($t(51)=2.82$, $MSE=2.16$, $p<.01$) however a significant interaction between variables emphasised this was not consistent across groups $F(6,96)=4.13$, $MSE=77.29$, $p<.01$.

Whilst all groups were more accurate matching whole faces than face parts, planned post hoc analyses investigated the difference between internal and external features. The CA group was more accurate using external than internal features ($t(12)=5.42$, $MSE=1.90$, $p<.01$), as was the NVMA group ($t(12)=3.86$, $MSE=3.19$, $p<.01$). Participants in the VMA group were also more accurate for external than internal features although the difference was not significant ($p=.32$). Participants with WS were more accurate using internal than external features $t(12)=3.57$, $MSE= 2.79$, $p<.01$.

Discussion

This paper assessed the ability of individuals with WS to match unfamiliar faces from whole faces and face parts (internal / external features). Previous research indicates that adults are more accurate recognizing familiar people from internal than external features, a pattern not evident for unfamiliar faces (e.g. Ellis et al., 1979). Importantly, the relative advantage for internal over external features is absent from unfamiliar face processing throughout development (e.g. Campbell et al., 1995, 1999). Research has never previously reported a relative advantage for internal features for unfamiliar faces with any population. This paper reports the first evidence of greater accuracy using internal features for unfamiliar face matching by individuals who have WS. Crucially, the same task produced the opposite results pattern for typically developing participants and therefore this pattern is unlikely to be driven purely by any effects of the specific stimuli being employed.

Performing at a level predicted by their verbal ability (against claims of ‘intact’ face processing e.g. Bellugi, Wang & Jernigan, 1994), participants with WS matched unfamiliar faces in a style previously cited only for familiar faces. Emphasising the overall cognitive profile associated with WS, performance was more accurate across all conditions than that of the nonverbal comparison group and demonstrates the superior performance on face tasks in comparison to other nonverbal abilities (e.g. see Bellugi et al., 2000).

The overall level of task performance on internal and external feature trials allowed group differences to be evident in a manner that was not possible in previous research using this paradigm (Deruelle et al., 2003). The crucial comparison here exposed the differential pattern of performance across typically and atypically developing participants on the same task. The number of trials in each condition may appear relatively low (to accommodate attentional requirements of participants) but importantly the same task produced a different results pattern across groups. Atypical unfamiliar face processing may directly relate to the social phenotype of WS; hypersociability and approachability towards strangers (e.g. Jones et al., 2000; Doyle et al., 2004), an interest in looking at faces irrespective of familiarity (Mervis et al., 2003) and extended face-gaze linked to lower physiological arousal (Doherty-Sneddon, et al., submitted). Research has also shown that individuals with WS are able to interpret cues from the upper internal face region, specifically the eyes, in a manner that dissociates individuals with WS from those with autism (Riby, Doherty-Sneddon & Bruce, accepted). The social phenotype of WS therefore appears to relate to aspects of face perception as assessed experimentally.

The results presented here may reveal not only aspects of the WS social phenotype, but aspects of the visual perception in WS. The current task splits and crops face regions into internal and external parts for the purposes of this assessment and may appear relatively unusual in appearance. The findings may therefore reveal less about face processing per se and may simply reflect atypicalities relating to the discrimination of separable parts or indeed linking together separable parts of an image. It is well documented that individuals with WS experience difficulties with grouping information and processing configurations on tasks such as the block design or when drawing (e.g. Farran, Jarrold, & Gatherole, 2003) and this may be extended to face processing tasks that unnaturally manipulate facial appearance. However, it has been well documented that linking together parts of a drawing (for example using a whole configuration) is possible when displays do not include biological information (e.g. Pani, Mervis, & Robinson, 1999). Care is therefore required when designing assessments for groups who may show atypicalities of visual perception.

In general, the results have implications for understanding typical face perception. Previous research emphasised that face familiarity affects the use of internal and external features. Evidence from WS provides possible suggestions to the underlying nature of this dissociation. Based on the social WS phenotype, a typical shift towards internal features for familiar faces may be more likely in social than informational terms. It is therefore hypothesised that there is an internal feature advantage for typical familiar (and not unfamiliar) face processing not because people have ‘learned’ the internal features of familiar people, but rather they are willing to attend to the internal features for familiar but not unfamiliar people. Participants with WS could not have ‘learned’ the specific internal features of these unfamiliar faces (due to repeat exposure etc) but cope well with such features. It could be argued that individuals with WS, due to their high levels of sociability, overfriendliness with strangers and decreased levels of physiological arousal, are more willing to attend to the internal parts of the face. If this sociability allows relatively more fixation on the internal features than is typical, the relative use of external features may be decreased and therefore this group become less adept at using the external face features for feature matching or identity processing. Explicitly, any differences between groups on external feature trials may be directly linked to the processing of internal features due to a shift in viewing preference or priorities in WS. The current research therefore supports notions of atypical socialisation in WS as evident here through experimental face perception paradigms.

As noted in the Introduction section, an assessment of unfamiliar face processing in WS may inform theories and models of typical face perception. The Bruce and Young (1986) model proposes different pathways for processing familiar and unfamiliar faces which may be implicated in the present study. The current results might be explained by individuals with WS using the FRUs and PINs (normally reserved for familiar faces) irrespective of familiarity or using a compensatory mechanism to allow for their relatively poorer performance using external features. The need for flexibility within the model is therefore emphasised by evidence from WS. Alternatively, the processing dissociation may occur at a deeper level than is accommodated by the Bruce & Young model. Dubois et al. (1999) argue against the strict independence of routes using evidence from positron emission tomography. They note that at the neurophysiological level activation of similar brain regions occurs irrespective of face familiarity. They do comment however that amygdala activation is greater for unfamiliar than familiar faces and this may have implications for individuals with WS where evidence suggests atypical amygdala activation (Meyer-Lindenberg, Mervis & Berman, 2006) and structure (Reiss et al., 2004). Hypofunctioning of the amygdala may be linked to behavioural evidence from face perception and entwined with the hypersocial phenotype associated with WS. Therefore the current study not only informs us of atypical face processing in WS but addresses the typical relationship between familiar and unfamiliar face perception strategies.

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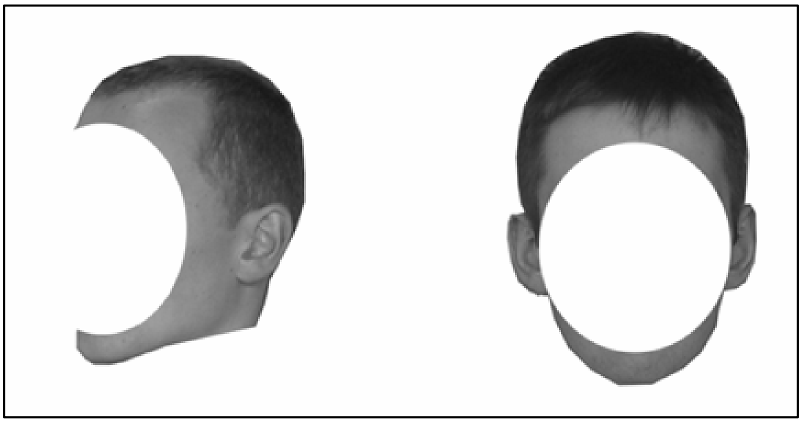
Figure Captions

Figure 1: An example of (i) an external feature different trial and (ii) an internal feature same trial.

Figure 2: Percentage correct for whole face, internal and external feature trials across groups

Figure 1

(i)



(ii)

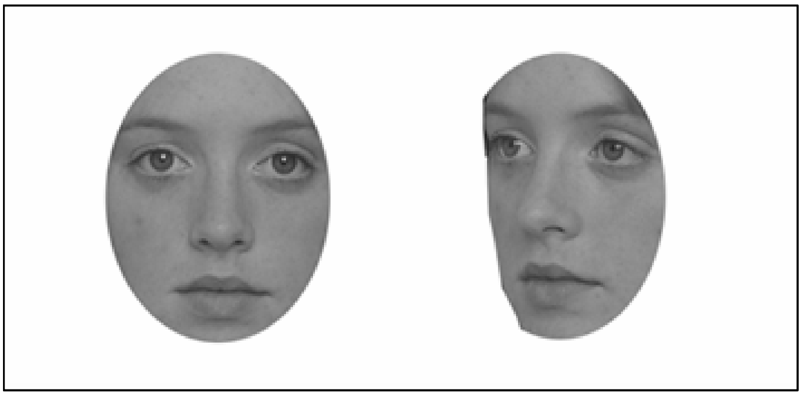
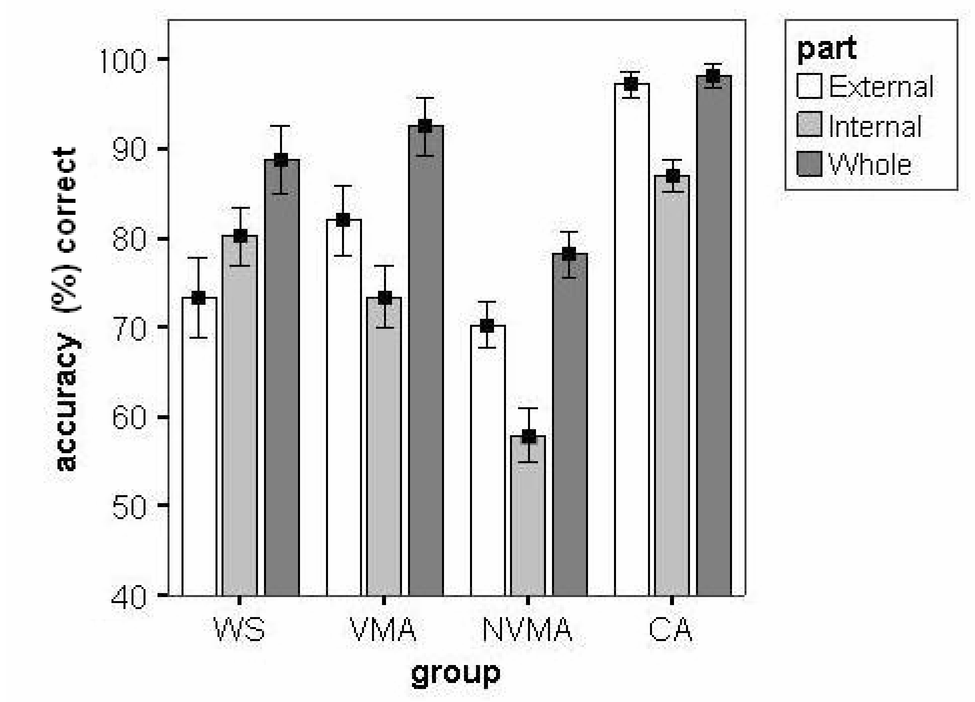


Figure 2



part
External
Internal
Whole
accuracy (%) correct
100
90
80
70
60
50
40
WS
VMA
NVMA
CA
group